

# Cloacal Exstrophy

## What is it?

Cloacal exstrophy, or **OEIS Syndrome**, is a rare birth defect. It is the most severe form in a group of birth defects called bladder exstrophy-epispadias complex (BEEC). Cloacal exstrophy occurs during the development of the abdominal wall structures before birth. As a result, an infant may be born with some abdominal organs exposed on the outside of the body. Pelvic bones, kidneys, and the spinal cord may also

be affected. Typically, the bladder is divided into two parts next to a section of the large intestine outside the body. In addition, boys are usually born with a flat, short penis that is separated into two halves. Girls are usually born with a clitoris that is separated into two halves. OEIS Syndrome consists of four distinct physical characteristics:

**Omphalocele:** Occurs when some organs (liver, spleen, intestine) push through the abdominal muscles near the umbilical cord and are exposed.

**Exstrophy of the rectum/bladder:** Rectum exstrophy occurs when the rectum is positioned outside of the body between the two bladder halves. Bladder exstrophy occurs when the bladder is divided into two halves outside of the abdomen and cannot hold urine.

**Imperforate anus:** Occurs when the anus is not formed or has no opening.

**Spinal defects:** Includes spina bifida, which is common in infants born with cloacal exstrophy.

## How common is it?

It is a rare and severe birth defect. It occurs in about 1 out of every 250,000 infants born each year.

## What causes it?

The cause of cloacal exstrophy is unknown. It could be caused by environmental and/or genetic factors.

## How is it diagnosed?

It is often diagnosed during pregnancy by a fetal ultrasound. Once the infant is born, the diagnosis can be confirmed by a physical exam.

## How is it treated?

Cloacal exstrophy is treated with reconstructive surgery that takes place throughout several years and includes different stages. Treatments include abdominal repair, osteotomies (bladder surgery), and pull-through surgery

(connects the colon to the rectum).

### For more information:

**Children's Hospital of Philadelphia**  
<http://www.chop.edu/conditions-diseases/bladder-exstrophy>

**Mayo Clinic**  
<https://www.mayoclinic.org/diseases-conditions/bladder-exstrophy/symptoms-causes/syc-20391299>

